PLG|D CONNECTIONS

BILATERAL LIGNEOUS CONJUNCTIVITIS WITH S. PNEUMONIAE SUPER-INFECTION IN A CHILD WITH TYPE I PLASMINOGEN DEFICIENCY

Literature-Based Case Study: 9-month-old female child (plasminogen activity, <5%)§

This patient first presented as an infant with ligneous conjunctivitis and infection, and experienced recurrence after topical treatment and surgical intervention. She was the daughter of consanguineous parents of Turkish background.

AGE

Physical examination
Bilateral swelling of both eyelids, hard white pseudomembranes on the upper and lower tarsal conjunctiva with partial lid eversion. On the left eye the bulbar conjunctiva was also involved.

Histological examination (pseudo-membrane)
Fibrin-rich with subepithelial deposition of amorphic hyaline material containing neutrophils and eosinophils

Initial diagnosis
Ligneous conjunctivitis with *Streptococcus pneumoniae* superinfection

Coagulation analysis lab results**
Plasminogen functional activity <5%
Plasminogen antigen: <0.4 mg/dl

Family testing
Both parents had low plasminogen activity but normal plasminogen antigen

Genetic testing (SSCP analysis)
No PLG gene genetic abnormality

Diagnosis
Severe Type I Plasminogen Deficiency (PLGD)

9 months

Inpatient initial treatment
Surgical excision of pseudomembranes and further treatment with topical heparin and erythromycin

Treatment results
Symptoms initially improved. Regrowth of pseudomembranes occurred 2 months postsurgery.

Inpatient follow up treatment
IV replacement therapy with lys-plasminogen* (daily infusion for 2.5 months)

IV lys-plasminogen* replacement results

- Plasminogen level began to increase within 30 min after infusion of lys-plasminogen (300–3000 casein units per 24 h) to normal values
- After 24 h the residual activity reached 20%
- Initial partial resolution of the conjunctival pseudomembranes. Stabilization of the ocular situation with no recurrence of pseudomembranes could be achieved for 6 months.

** IV Lys-plasminogen is not commercially available*

*** Normal levels of plasminogen activity: 70-130%¥*
Normal levels of plasminogen antigen: 6-25 mg/dl§

References:
§ Kraft et al. Graefes Arch Clin Exp Ophthalmol. 2000;238(9):797-800.
¥ Rare bleeding disorders. http://www.rarecoagulationdisorders.org/diseases/plasminogen-deficiency/laboratory-evaluation. Accessed April 21, 2017.
† Tefs et al. *Blood.* 2006;108(9):3021-3026. Based on a study of 50 patients with PLGD from 44 families.

KEY TAKEAWAYS

- Ligneous conjunctivitis is a hallmark manifestation of PLGD; approximately 80% of PLGD patients experience ligneous conjunctivitis†.
- If ligneous conjunctivitis is suspected, it is recommended to test for PLGD by measuring plasminogen activity and antigen level, and conducting genetic testing.
- PLGD is an ultra-rare genetic disease that can have devastating effects on multiple organ systems.